

CASE REPORT

Imaging of a fourth ventricle arachnoid cyst in a child

F. Lockhat^a, L. Mubaiwa^b, B. Clarke^c, P. Corr^{a,*}*Departments of ^aRadiology, ^bPaediatric Neurology, and ^cAnatomical Pathology, Nelson R Mandela Medical School, University of KwaZulu-Natal, South Africa*

Introduction

In the aetiology of obstructive hydrocephalus, arachnoid cysts of the fourth ventricle are a rare cause of obstructive hydrocephalus, with only a few cases described in the literature.^{1,2} We present the MR findings in the case of a child who presented with hydrocephalus due to an obstructive cystic mass lesion within the fourth ventricle; the lesion was histologically proven to be an arachnoid cyst.

Case report

A little girl aged 2.5 years presented with left-sided facial weakness of acute onset and approximately 9 months duration (according to the mother). There was no history of a previous viral illness, ear discharge or trauma. The child's behaviour was reported as normal and there was no impairment in developmental and motor skills. Her delivery had been normal and her birth weight was 2.1 kg.

Neurological examination confirmed a left 7th cranial nerve lower motor nerve palsy, with no associated 6th nerve involvement or bulbar palsy. Fundoscopy was normal and there were no signs of raised intracranial pressure, nor were cerebellar signs elicited. Power and deep tendon reflexes in the limbs were normal. CT demonstrated a cystic mass of the fourth ventricle, and MRI was requested for further evaluation.

MRI of the brain was performed before and after gadolinium DTPA contrast injection, in the sagittal

and axial planes, with pre-contrast T1 and T2 and post-contrast T1 sequences (Fig. 1(a) and (b)), FLAIR and diffusion-weighted images (Fig. 1(c) and (d)). A non-enhancing cystic mass within the fourth ventricle was demonstrated. Diffusion imaging was helpful in depicting normal unrestricted diffusion, so confirming that the lesion was most likely an arachnoid cyst: the main differential diagnoses of a fourth ventricular lesion are arachnoid cyst, neuroepithelial cyst and epidermoid tumour.

After surgical excision, histological examination confirmed an arachnoid cyst consisting of fibrous connective tissue lined by a thin layer of meningotheial cells.

Discussion

Arachnoid cysts of the fourth ventricle are congenital and rare, comprising 1% of all intracranial masses¹⁻⁴ and appearing as intra-arachnoid space-occupying lesions; 75% of these occur in children, with a male to female ratio of 3:1.⁵ The clear fluid within the cysts is identical to CSF. The aetiology is related to maldevelopment of the meninges caused by an aberration in CSF flow through loose embryonic perimedullary mesenchyme.⁵ This results in focal splitting of developmental meninges, leading to the formation of a diverticulum between the arachnoid and pia.⁵

Clinically, arachnoid cysts present as small, mostly asymptomatic lesions.⁶ Those involving the middle cranial fossa are associated with temporal lobe hypogenesis and often cause erosion and expansion of the overlying cranium.³ A large lesion may be associated with mass effect and compressive symptoms, with raised intracranial pressure resulting in hydrocephalus.⁶ Arachnoid cysts are often supratentorial in location: 50 to 65% are found

* Guarantor and correspondent: P. Corr, Department of Radiology, Nelson Mandela Medical School, University of Natal, Private Bag 7, Congella, 4013 Durban, South Africa. Tel.: +2731 260 4301; fax: +2731 260 4621.

E-mail address: corr@nu.ac.za (P. Corr).

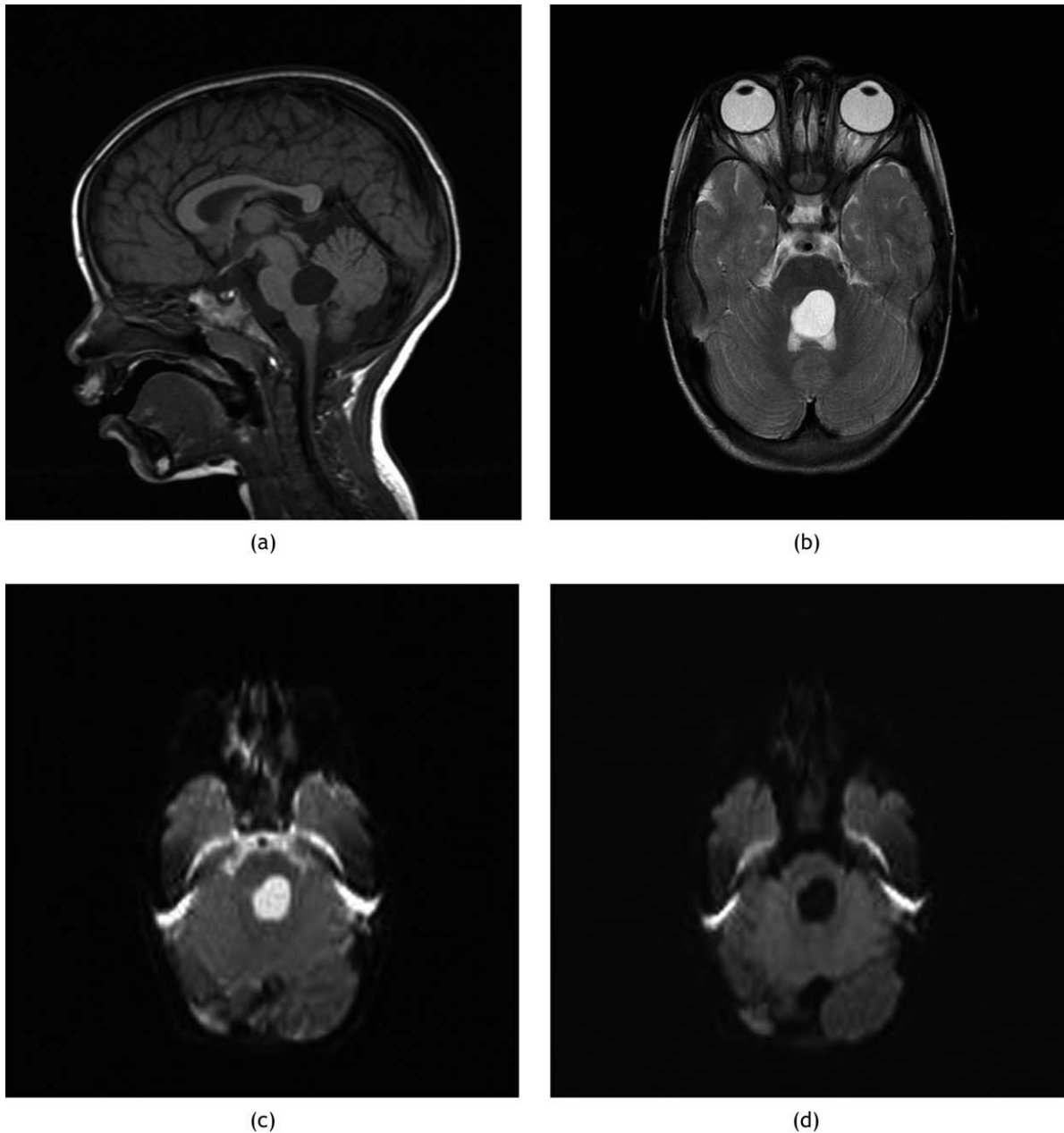


Figure 1 (a) Sagittal T1-weighted MR image demonstrates a hypointense arachnoid cyst compressing the fourth ventricle and cerebellum with the same signal intensity as CSF. (b) Axial T2-weighted MR features a hyperintense arachnoid cyst in the floor of the fourth ventricle, compressing the ventricle posteriorly. Note that the signal intensity is identical to that of CSF. (c) Medium diffusion-weighted axial image (b0 400) through the cyst shows signal hyperintensity identical to CSF. (d) High diffusion-weighted axial image (b0 1000) through the cyst exhibits signal hypointensity identical to CSF, in keeping with unrestricted diffusion.

in the middle cranial fossa. Less frequent locations include the cerebral convexities, cerebropontine angles, retro-vermian space and the quadrigeminal plate cisterns, suprasellar cistern (5 to 10%) and cisterna magna.^{5,6}

On CT, an arachnoid cyst appears as a smooth, non-calcified, extra-axial, non-enhancing lesion with a density identical to that of CSF.⁵

On MRI, the lesion appears sharply demarcated and extra-axial; it displaces or deforms adjacent brain. The cyst has no internal architecture and does not demonstrate significant contrast enhancement.⁵ Hyperintensity is seen on long TR/TE images and hypointensity on short TR/TE images, or the appearance may be isointense on long TR/TE images.³ Signal intensity is identical to that of

CSF, but protein in the cyst fluid may lead to T1 hyperintensity. Haemorrhage also may increase the signal intensity on all sequences.⁶

Arachnoid cysts demonstrate low signal intensity on diffusion-weighted images, which is identical to the appearance of CSF on diffusion-weighted and FLAIR imaging.^{7,8} However, both arachnoid cysts and CSF can be differentiated from epidermoid cysts by the abnormal, higher signal intensity of the latter on diffusion-weighted and FLAIR imaging.^{4,7}

Pathologically, arachnoid cysts have a thin transparent wall separated from inner dura or pia. Microscopically, the cyst wall consists of a vascular and collagenous membrane lined by flattened arachnoid cells. No glial limiting membrane or epithelial lining is seen.⁵ The wall consists of elongated epithelial cells and connective tissue with lamellar collagen fibre bundles.⁹ The columnar epithelial cells stain positive with the PAS reagent.⁹

Treatment of symptomatic cysts consists of decompression of the cyst by shunting the fluid, rather than excision.⁶

Conclusion

Paediatric arachnoid cysts of the fourth ventricle are rare. This case report describes the invaluable

role of MRI in establishing the diagnosis. MRI assists in differentiating between other lesions of the fourth ventricle, using specialized techniques such as diffusion imaging.

References

1. Makiuchi T, Kadota K, Asakura T, et al. Arachnoid cyst of the fourth ventricle. *Neurol Med Chir* 1990;30:848–52.
2. Erdinder P, Kaynar MY, Boskus H, Ciplak N. Posterior fossa arachnoid cysts. *Br J Neurosurg* 1999;13:10–17.
3. Scott WA, editor. *Magnetic resonance imaging of the brain and spine*. 1st ed. New York: Raven Press; 1991. p. 364–8.
4. Stark DD, Bradley Jr WG. *Magnetic resonance imaging*. 3rd ed. St Louis, MI: Mosby; 1999 [p. 1421-1422, 1476-1478].
5. Osborne AG. *Diagnostic neuroradiology*. 1st ed. St Louis, MI: Mosby; 1994 [p. 65-66, 639-642].
6. Sartor K. *MR imaging of the skull and brain*. 1st ed. Berlin: Springer; 1992 [p. 187-195].
7. Fischbein NJ, Dillon WP, Barkovich AJ. *Teaching atlas of brain imaging*. 1st ed. New York: Thieme Medical Publishers; 2000 [79-81].
8. Yamaguchi S, Hirohata T, Sumida M, Arita K, Kurisu K. Intradiploic arachnoid cyst identification by diffusion weighted magnetic resonance imaging—case report. *Neurol Med Chir* 2002;42:137–9.
9. Miyagami M, Kasahara E, Miyazaki S, Tsubokawa T, Kagawa T. Ultrastructural findings of arachnoid cysts and epithelial cysts. *No to Shinkei* 1991;43:545–53.